Quarterly Shareholder Update – June 2020

Dear Shareholder,

The March Quarter saw Pharmaxis making rapid adjustments required by the outbreak of COVID-19 including ensuring continuity of supply for patients receiving Bronchitol and changed workplace arrangements for our employees.

Despite these changes, I am pleased to report that the June Quarter has been very productive with some important market announcements such as the filing of an investigational new drug (IND) application for our myelofibrosis drug along with a significant amount of activity going on in the background to reshape the Pharmaxis business for the future.

First a comment on announcements since the last Quarterly Update that foretell of pivotal H2 2020 events.

- **IND filing for our myelofibrosis drug; PXS-5505**
  Unlike the majority of IND filings that seek approval for the first trial in humans, Pharmaxis has already completed phase 1 studies in healthy volunteers so the IND is a much more substantial document containing over 20,000 pages of reports on both pre-clinical and clinical testing to support approval for a phase 2 study in the rare bone cancer myelofibrosis. We are already in discussions with potential trial sites and, subject to FDA approval, anticipate the start of recruitment to be in Q4 2020.
- **Bronchitol US FDA review**
  Our US licensee Chiesi filed its updated response to the US Food and Drug Administration (FDA) in the June quarter and the FDA advised it will complete its review by 1 November 2020. This is a little later than Chiesi had expected but both Chiesi and Pharmaxis are working towards a US Bronchitol launch in Q1 2021 with a US$10m milestone payment due to Pharmaxis at that time.

There are two significant additional work streams linked to these events that we are pursuing:

- **Refocusing the business**
  The development and commercialisation of Bronchitol reaches a pivotal point on November 1st. Approval by the FDA for Bronchitol would see the mannitol business segment generate immediate cash and move into profitability. I recognise that the immediate returns may not be transformative, but this development does provide an opportunity to investigate different ways of structuring our business and funding the drug development activities that drive shareholder value in a much more significant way. Pharmaxis and our partners are already in discussions about how to shape the future post November 1st.
- **Accelerating drug development**
  The data in the IND submission for PXS-5505 includes long term toxicity studies, human PK and PD data and GMP manufacture of clinical trial supplies. In short, it's a launching pad to phase 2 clinical proof of concept studies that will attract the attention of patients and physicians and should also excite investors. Our priority is myelofibrosis, but this drug has potential in a number of other cancers such as pancreatic cancer, oral cancer, and glioblastoma. We have been busy building collaborations with academic and clinical centres worldwide who have welcomed the prospect of PXS-5505 for their patients and the IND will open the door to explore investigator initiated, grant funded trials in a number of these diseases.

I expect the rest of the year to be busy with announcements on LOXL2 partnering, Boehringer's decision on continuing development in diabetic retinopathy, and other milestones and events for the Pharmaxis early stage pipeline all pending.

Sincerely,

Gary Phillips - Chief Executive Officer

Products and Pipeline at a glance

Disease/condition	Drug	Status
Cystic fibrosis	Bronchitol	Approved
Asthma	Aridol	Approved
AOC3 inhibitor	BI1467335	Partnered Phase 2
LOXL2 inhibitor	PXS-5382; PXS-5338	Phase 1 completed
Systemic pan-LOX inhibitor	PXS-5505	Phase 2 ready
Topical pan-LOX inhibitor	PXS-6302	Pre-clinical

Impact of COVID-19

Pharmaxis' initial response to the COVID-19 global pandemic was outlined in the March shareholder update, describing the precautions the Company was taking to protect employees and to continue manufacturing and supply of its approved respiratory products.

As reported there was no impact of COVID-19 on the various clinical trials in which the Company is involved. Furthermore, the Company has continued an uninterrupted supply to local and global customers, despite there being significantly fewer international freight routes.

As further discussed below there was a reduction of Aridol sales in particular in April and May.

While employees are now able to spend more days in the office and labs with social distancing and enhanced cleaning protocols the default of working from home remains in place for those who can.

The Company continues to monitor the situation.

Drug discovery

Boehringer Ingelheim development of BI 1467335

BI 1467335 (formerly known as PXS-4728A) was acquired by Boehringer Ingelheim in 2015 with an upfront payment to Pharmaxis of $41 million.

Boehringer initiated a phase 2a proof of clinical principle trial for the eye disease diabetic retinopathy (DR) in January 2018.

Under the terms of the agreement Boehringer has total responsibility for the development program. Pharmaxis has received a total of A$42 million in development milestones to date from Boehringer, bringing the total received to $83 million.

The phase 2a study of the drug in DR was completed in the June quarter and will report in H2 of 2020. Future development will be determined by Boehringer in H2 2020 following review of the phase 2a study report.

Pharmaxis receives payments upon achievement of certain development milestones. Future potential payments by Boehringer to Pharmaxis for the DR program include:

- Commencement of phase 3 clinical trial milestone: €37m (approximately A$62m)
- Filing, approval and pricing milestones: total of €140m (approximately A$230m)
- Sales related payments increasing from high single digits
- Sales milestones

Diabetic retinopathy is the leading cause of vision-loss in adults. Of an estimated 285 million people with diabetes mellitus worldwide, approximately one third have signs of DR and of these, a further one third is vision-threatening.

LOXL2 inhibitor program

The Lysyl Oxidase Like 2 (LOXL2) enzyme is fundamental to the fibrotic cascade that follows chronic inflammation in the liver disease NASH, cardiac fibrosis, kidney fibrosis and idiopathic pulmonary fibrosis (IPF) and it also plays a role in some cancers.

The Pharmaxis drug discovery group developed two small molecule inhibitors to the LOXL2 enzyme that have completed phase 1 clinical trials

and 3-month toxicology studies (PXS-5382 and PXS-5338).

During the quarter the Company completed an additional small phase 1 study in response to questions from pharma companies interested in the program, demonstrating an improved pharmacokinetic profile in a number of different dosing regimens.

Pharmaxis is currently pursuing a number of different partnering options to enable the drug to enter the clinic in phase 2 trials. While the process has taken longer than originally expected, and much of the industry focus has been on the Covid-19 pandemic, the Company continues to have discussions with a number of potential partners. Pharmaxis will provide more information when the process concludes.

Systemic LOX inhibitor program

Pharmaxis is progressing two lysyl oxidase (LOX) programs from its amine oxidase chemistry platform, both of which are planned to be partnered at a later stage of development - after phase 2 clinical trials.

The most advanced LOX program has developed an oral drug (PXS-5505) that inhibits all lysyl oxidase family members (LOX, LOXL1, 2, 3 & 4). The compound successfully cleared pre-clinical safety including 6-month toxicity and has shown significant reductions in fibrosis in in-vivo models of myelofibrosis (MF), pancreatic cancer and other cancers as well as in models of systemic sclerosis. PXS-5505 has shown to be well tolerated in Phase 1 single and multiple ascending dose studies that reported earlier in the year with an excellent pharmacokinetic and pharmacodynamic profile.

Based on pre IND feedback from the US Food and Drug Administration (FDA) on the PXS-5505 program in MF and discussions of the trial protocol with key opinion leaders in the US, Europe and Australia, the Company has recently filed an investigational new drug (IND) application with the FDA. The FDA takes 30 days to review IND applications.

Shortly after the end of the quarter the FDA granted orphan-drug designation for the pan LOX inhibitor PXS-5505 for the treatment of myelofibrosis.

Pharmaxis has appointed an international clinical research organisation to manage the phase 1/2 clinical trial which is scheduled to commence in Q4 2020, subject to the FDA review of the IND. Parexel has identified sites where the current COVID-19 pandemic will not prevent recruitment.

Pharmaxis also recently received orphan drug status from the FDA for this program.

Topical LOX inhibitor program

The Company's other LOX program has developed a drug for topical application with the potential for use in scar revision, keloid scarring and scarring from burn wounds.

A lead candidate has been selected (PXS-6302) and completed pre-clinical development including initial stability studies of the topical formulation.

The Company has ongoing discussions with an Australian based hospital burns units that is interested in commencing a series of investigator initiated clinical studies to assess the safety and initial efficacy of this drug in burns related scars and pre-existing scars. The studies are currently scheduled to commence in the second half of the 2020 calendar year.

Mannitol business

Bronchitol and Aridol

Bronchitol®(mannitol) is an inhaled dry powder for the treatment of cystic fibrosis (CF) and has been the subject of three large scale global clinical trials conducted by Pharmaxis. The product is approved and marketed in Australia, Europe, Russia and several other countries.

Aridol® is an innovative lung function test designed to help doctors diagnose and manage asthma. Aridol is approved for sale in Australia, major European countries, the United States, Canada and South Korea.

United States

The Company's US partner Chiesi Group is responsible for the commercialisation of Bronchitol in the United States.

Following receipt of a complete response letter from the FDA in June 2019 and subsequent discussions with the FDA, Chiesi has revised its

product packaging and user instructions for Bronchitol and completed the human factor study (HFS) required to demonstrate that healthcare professionals can properly administer the mannitol tolerance test – an initial test to ensure patients hypersensitive to mannitol are not prescribed Bronchitol. Chiesi filed its updated response to the FDA in the current quarter and, in acknowledging receipt of the filing the FDA advised it will complete its review by 1 November 2020.

Subject to approval, Pharmaxis will receive a US$10 million milestone payment on the commercial launch of Bronchitol in the US, mid to high teen percentage royalties and will be the exclusive supplier of Bronchitol for the US market.

Western Europe

In the EU, Chiesi is the Pharmaxis exclusive Bronchitol distributor for the markets of the UK, Ireland, Germany, Italy, Norway, Sweden, Finland, Denmark, Cyprus and Greece.

Pharmaxis also markets Bronchitol in Austria via its German based logistics provider and in Switzerland via an exclusive distributor.

Other territories

Bronchitol is sold in Australia by Pharmaxis and in Turkey, the Czech Republic, Hungary and Russia by exclusive distributors.

Bronchitol sales

Bronchitol sales for the quarter and twelve months ended 30 June 2020 and 30 June 2019 are as follows:

$'000	Three months		Twelve months	
	2020	2019	2020	2019
Australia	227	277	1,221	1,059
Western Europe	1,613	24	2,638	1,041
Russia & Eastern Europe	624	306	1,403	464
Total	**$2,464**	**$607**	**$5,262**	**$2,564**

Pharmaxis ex-factory sales for the current quarter reflect the buying patterns of its international distributors where large orders were shipped to both our EU and Russian distributors. Unit in-market sales of Bronchitol by Chiesi in the UK, Germany and Italy for the three and twelve months ended 30 June 2020 increased 8% over 2019.

Pharmaxis Bronchitol distributors typically order on a six monthly basis.

The COVID-19 global pandemic has not to date impacted purchasing of Bronchitol by our international distributors. In Australia where Pharmaxis sells directly to clinics, unit sales were 18% lower than the prior quarter, although unit sales from the start of the pandemic in March to June were marginally above the same period in 2019, possibly indicating clinics purchasing ahead of expected ordering difficulties. The Company continues to monitor the situation.

Aridol

At the beginning of the COVID-19 pandemic a number of countries, including those where Aridol is sold, respiratory specialists were advised to limit all lung function testing to more severe cases due to health risks arising from patients exhaling multiple times with force as part of the test. In the markets where Pharmaxis sells Aridol directly to lung function testing laboratories (Australia and Europe) average monthly sales for the period 1 March to 30 June 2020 were $63,000 per month, compared to $126,000 per month in the same period in 2019. April and May were the months most impacted with June 2020 sales increasing to 69% of June 2019. We have been advised that the limitation on lung function testing has been eased in a number of markets. The Company continues to monitor the situation.

Aridol sales

Aridol sales for the quarter and twelve months ended 30 June 2020 and 30 June 2019 are as below.

$'000	Three months		Twelve months	
	2020	2019	2020	2019
Australia	59	131	436	471
Europe	71	246	912	979
USA & Canada		344	72	1003
South Korea	87	177	345	659
Total	$217	$898	$1,765	$3,112

Following two large orders in the previous financial year, there were no US Aridol orders during the half from the Company's North American distributor.

Corporate

Dr Neil Graham Appointed as Director

On 4 May Pharmaxis announced the appointment of experienced senior global pharma and biotech executive Dr Neil Graham to the Board of Directors.

Dr Graham is a medicines development expert and infectious diseases epidemiologist who has had a distinguished academic and business career. He recently joined Evelo Biosciences (EVLO:US) as Chief Development Officer after eleven years as Vice President of Strategic Program Direction at Regeneron (REGN:US) where he was responsible for the pipeline portfolio and successfully leading development of immunology and inflammation products from preclinical development to post-launch.

Between 2007 and 2009 he was Senior Vice President, Program and Portfolio Management at Vertex, Cambridge, USA, overseeing all development programs including studies in cystic fibrosis and inflammation. He has previously led clinical development programs as a senior executive at Trimeris Inc. and XTL Biopharmaceuticals.

Dr Graham was educated at University of Adelaide (MBBS, MD, MPH). Between 1993 and 1997 he was Associate Professor of Epidemiology at John Hopkins University School of Hygiene and Public Health with research focused on HIV, tuberculosis and hepatitis. He has authored a number of books and publications including a considerable body of work on respiratory illness.

Dr Graham's global career in drug development is directly relevant to the Company's current development assets such as the systemic pan-LOX inhibitor that will move into phase 2 studies later this year.

Dr Graham is based in the United States.

2020 Annual General Meeting

The 2020 Annual General Meeting of Pharmaxis Ltd will be a virtual meeting, and will be conducted online at 10.00am on 4 November 2020. Video and online technology will be used to facilitate shareholder engagement and participation in the meeting. Details of where shareholders can access the notice of meeting, lodge a proxy and participate in the meeting will be sent to each shareholder. If you choose to participate online on the day of the meeting you will be able to view a live webcast of the meeting, ask the Directors questions and submit your votes in real time.

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Financials

Key financial metrics

A$'000	Three months ended		Twelve months ended	
(unaudited)	**30-June-20**	**30-June-19**	**30-June-20**	**30-June-19**
Income statements				
Sales of Bronchitol & Aridol	2,681	1,505	7,027	5,676
Total revenue	7,725	7,797	13,029	13,080
Total expenses	(1,810)	(8,939)	(26,972)	(33,138)
Net profit (loss) after tax	**5,915**	**(1,142)**	**(13,943)**	**(20,058)**
Segment results – adjusted EBITDA				
Mannitol business	(244)	(1,819)	(3,977)	(5,013)
New drug development	2,011	2,462	(5,113)	(6,764)
Corporate	(707)	(1,335)	(2,990)	(3,874)
Total	**1,060**	**(692)**	**(12,080)**	**(15,651)**
Statement of cash flows				
Cash inflow/ (outflow) from:				
Operations	(4,729)	(3,404)	(13,284)	(19,798)
Investing activities	(116)	(124)	(574)	(981)
Financing activities	(642)	(477)	(2,502)	20,830
Total cash generated/(used)	**(5,487)**	**(4,005)**	**(16,360)**	**51**
Cash at bank	**14,764**	**31,124**	**14,764**	**31,124**

Highlights

- Revenue
  - See above for detail and commentary on Bronchitol and Aridol sales which increased 78% for the quarter and 24% for the full year over the prior period.
  - The Company booked an R&D tax incentive of $4.9 million in the quarter ($5.2 million for the full year).
- Expenses
  - When unrealised foreign exchange rate gains and losses and other adjustments in relation to the NovaQuest financing agreement are excluded, total expenses for the quarter were $7.4 million (2019: $8.7 million) and for the full year $29.1 million (2019: $32.0 million). This reduction in expenses compared to the prior quarter and nine months is primarily due to lower drug discovery expenditure and positive changes in inventory offset by increased clinical trial costs and depreciation and amortisation.
  - Finance costs were a credit of $2.6 million for the quarter (2019: expense of $0.1 million) and a credit of $2.2 million for the full year (2019: expense of $0.2 million). This included a $2.7 million reduction in the financing liability subsequent to a review of the estimated cash flows expected to be paid under the agreement.

- Net loss
  - Net loss for the full year reduced from $20.1 million in 2019 to $13.9 million in 2020.
- Cash
  - The Company finished the quarter with $14 million in cash.
  - Operational cash flows for the nine months include receipt of the 2019 R&D tax incentive of $6.2 million. The Company expects to receive its 2020 R&D tax incentive of $4.9 million in the second half of the 2020 calendar year.

Segment information

A$'000	Segment information - three months ended							
(unaudited)	30-June-20				30-June-19			
Income statements	Mannitol Business	New Drug Development	Corporate	Total	Mannitol Business	New Drug Development	Corporate	Total
Revenue								
Sale of Bronchitol	2,464			2,464	607			607
Sale of Aridol	217			217	898			898
	2,681			**2,681**	1,505			**1,505**
Tax credit		4,900		4,900		5,962		5,962
Other revenue	5		92	97	7		128	135
	2,686	**4,900**	**92**	**7,678**	1,512	**5,962**	**128**	**7,602**
Expenses								
Employee costs	(1,302)	(901)	(344)	(2,547)	(1,636)	(671)	(433)	(2,740)
Clinical trials		(539)		(539)		(1,118)		(1,118)
Drug discovery		(1,342)		(1,342)		(1,516)		(1,516)
Changes in inventories	(631)			(631)	(579)			(579)
Other expenses	(997)	(107)	(455)	(1,559)	(1,116)	(195)	(524)	(1,835)
Total expenses	(2,930)	(2,889)	(799)	(6,618)	(3,331)	(3,500)	(957)	(7,788)
Adjusted EBITDA	**($244)**	**$2,011**	**($707)**	**$1,060**	**($1,819)**	**$2,462**	**($829)**	**($186)**

Commentary for the quarter

- Mannitol Business:
  - Sales of Bronchitol and Aridol are detailed and discussed in the commentary above.
  - Increased revenue of $1.1 million combined with unchanged expenses resulted in Adjusted EBITDA improving from a loss of $1.8 million in 2019 to a loss of $0.2 million in 2020.
  - The reduction in employee expenses for the quarter reflects redeployment of clinical and regulatory resources to New Drug Development in support of the upcoming clinical program in myelofibrosis.
- New Drug Development:
  - The increase in employee expenses reflects redeployment of clinical and regulatory resources from the Mannitol Business.
  - Clinical trial expenses include the phase 1 trial for the Systemic LOX program of $211,000 (2019: $1.1 million) and a small phase 1 dosing study in LOXL2 of $329,000 (2019: $6,000).
  - Drug discovery expenses include work on the Topical LOX topical program ($369,000 for the quarter; $512,000 in 2019); work on the Systemic LOX program ($565,000 for the quarter; $411,000 in 2019); and work on the SSAO combo programs ($156,000 for the quarter; $256,000 in 2019).

- Corporate
  - The decrease in employee expenses reflects the cancellation of senior management bonuses for the year.
  - The decrease in other expenses reflects a reallocation of occupancy costs effective from the beginning of the current financial year.

A$'000	Segment information - twelve months ended							
(unaudited)	**30-June-20**				**30-June-19**			
Income statements	Mannitol Business	New Drug Development	Corporate	Total	Mannitol Business	New Drug Development	Corporate	Total
Revenue								
Sale of Bronchitol	5,262			5,262	2,564			2,564
Sale of Aridol	1,765			1,765	3,112			3,112
	7,027			**7,027**	**5,676**			**5,676**
Tax credit		5,159		5,159		5,962		5,962
Other revenue	20		459	479	27		506	533
	7,047	**5,159**	**459**	**12,665**	**5,703**	**5,962**	**506**	**12,171**
Expenses								
Employee costs	(5,855)	(3,373)	(1,637)	(10,865)	(6,083)	(2,837)	(1,932)	(10,852)
Clinical trials	98	(2,730)		(2,632)	621	(2,975)		(2,354)
Drug discovery		(3,709)		(3,709)		(6,308)		(6,308)
Changes in inventories	(1,457)			(1,457)	(1,689)			(1,689)
Other expenses	(3,810)	(460)	(1,812)	(6,082)	(3,565)	(606)	(2,448)	(6,619)
Total expenses	(11,024)	(10,272)	(3,449)	(24,745)	(10,716)	(12,726)	(4,380)	(27,822)
Adjusted EBITDA	**($3,977)**	**($5,113)**	**($2,990)**	**($12,080)**	**($5,013)**	**($6,764)**	**($3,874)**	**($15,651)**

Commentary for the twelve months

- Mannitol Business:
  - Sales of Bronchitol and Aridol are detailed and discussed in the commentary above, increasing 24% over the prior year.
  - Adjusted EBITDA reduced from a loss of $5 million in 2019 to $4 million in 2020.
  - The reduction in employee expenses reflects redeployment of clinical and regulatory resources to New Drug Development in support of the upcoming clinical program in myelofibrosis.
  - Clinical trial credits were received in both 2020 and 2019 from the contract research organisation that managed the clinical trial CF303.
  - The increase in other expenses reflects higher distribution costs in the EU and one-off regulatory and pharmacovigilance consultant costs.
- New drug development:
  - The increase in employee expenses reflects redeployment of clinical and regulatory resources from the Mannitol Business.
  - Clinical trial expenses for 2020 include the phase 1 trial for the Systemic LOX program of $1.8 million and a small phase 1 dosing study in LOXL2 of $935,000. In 2019, the clinical trial expenses related to the phase 1 trial for the Systemic LOX program of $1.6 million and the main phase 1 trials conducted in the LOXL2 program ($1.4 million).
  - Drug discovery expenses include work on the Topical LOX topical program ($1.2 million; $1.5 million in 2019); the Systemic LOX program ($1.3 million; $2.2 million in 2019); and the SSAO

combo programs ($0.5 million; $1.2 million in 2019). Prior period expenses included work on the LOXL2 program ($1.0 million.

- Corporate
  - The decrease in other expenses includes a reallocation of occupancy costs effective from the beginning of the current financial year.

Authorised for release to the ASX by Pharmaxis Ltd Disclosure Committee.
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Income statements

A$'000	Three months ended		Twelve months ended	
(unaudited)	**30-June-20**	**30-June-19**	**30-June-20**	**30-June-19**
Revenue				
Revenue from sale of goods	2,681	1,505	7,027	5,676
Interest	48	195	364	909
R&D tax incentive	4,900	5,962	5,159	5,962
Other	96	135	479	533
Total revenue	**$7,725**	**$7,797**	**$13,029**	**$13,080**
Expenses				
Employee costs	(2,380)	(2,854)	(11,425)	(11,928)
Administration & corporate	(432)	(492)	(2,041)	(2,179)
Rent, occupancy & utilities	(253)	(348)	(999)	(1,386)
Clinical trials	(539)	(1,119)	(2,632)	(2,354)
Drug development	(1,342)	(1,516)	(3,709)	(6,308)
Sales, marketing & distribution	(291)	(375)	(1,346)	(1,136)
Safety, medical and regulatory affairs	(383)	(280)	(1,058)	(896)
Purchases and changes in inventories	(630)	(579)	(1,456)	(1,689)
Other	(90)	(373)	(592)	(1,065)
Depreciation & amortisation	(812)	(665)	(3,236)	(2,619)
Foreign currency exchange gains & losses	2,732	(225)	(638)	(1,340)
Finance costs	2,610	(113)	2,160	(238)
Total expenses	**(1,810)**	**(8,939)**	**(26,972)**	**(33,138)**
Net profit (loss) before tax	**5,915**	**(1,142)**	**(13,943)**	**(20,058)**
Income tax expense				
Net profit (loss) after tax	**$5,915**	**($1,142)**	**($13,943)**	**($20,058)**

Summary balance sheets

A$'000 (unaudited)	30-June-20	30-June-19
Assets		
Cash	14,764	31,124
R&D tax incentive	4,900	5,962
Accounts receivable	1,459	1,171
Inventory	2,630	2,116
PP&E	8,906	9,123
Other	2,757	2,032
	$35,416	**$51,528**
Liabilities		
Accounts payable and accrued expenses	2,765	4,194
Lease liability (Frenchs Forest facility)	8,154	7,171
Financing agreement (not repayable other than as a % of US & EU Bronchitol revenue)	21,200	23,626
Other liabilities	1,866	1,723
	$33,985	**$36,714**